Primary Nonessential Cutis Verticis Gyrata

Cutis verticis gyrata primaria no esencial

To the Editor:

A 16-year-old boy with mental retardation was referred to our department because of folds in the scalp that had begun to develop 10 years earlier. The patient had no family history of similar lesions and denied any previous history of inflammation of the scalp or signs and symptoms of neurologic or psychiatric disorders.

Physical examination revealed folds and furrows running in an anteroposterior direction over the scalp that could not be corrected by traction (Fig. 1). No areas of alopecia were observed nor were there differences in hair distribution between affected areas and areas of normal skin. The rest of the physical examination revealed no other significant skin lesions.

A full laboratory workup, including a complete blood count, biochemistry, urinalysis, magnetic resonance imaging, skin biopsy, and chromosome analysis revealed no significant alterations, and there were no relevant findings on ophthalmologic study.

Based on the clinical features and the results of the tests performed, we made a diagnosis of primary nonessential cutis verticis gyrata. The patient and his family were informed of the benign nature of the lesion and no treatment was performed. Subsequent follow-up revealed no change in the condition.
Cutis verticis gyrata, first described in 1837 by Alibert, is a term that refers to a condition characterized by folds or deep furrows that resemble the surface of the brain.\(^1\) The male to female ratio is 5-6 to 1. Presentation is before puberty in the majority of cases and only 10% develop after 30 years of age; secondary cutis verticis gyrata can appear at any age, depending on the underlying pathology. The etiology of this condition is varied and 2 groups have been described, with primary and secondary variants. Cutis verticis gyrata may thus be classified as primary (essential and nonessential) or secondary. The term primary essential is used in cases with no other associated abnormalities, and primary nonessential refers to cases associated with mental retardation, cerebral palsy, epilepsy, schizophrenic, neurologic abnormalities, deafness, ophthalmologic abnormalities, or a combination of any of these. Secondary cutis verticis gyrata has been associated with various other conditions, including inflammatory diseases, hamartomatous lesions, tumors, acromegaly, myxedema, idiopathic hypertrophic osteopathy, amyloidosis, syphilis, leukemia, cretinism, acanthosis nigricans, tuberous sclerosis, neurofibromatosis, Ehlers-Danlos syndrome, and trauma.\(^2-5\) Chromosome abnormalities have been observed in some cases.

Although the etiology and pathogenesis are unknown, autosomal recessive and dominant factors have been detected in sporadic cases of the primary essential form, and endocrine disturbances could be implicated in the primary nonessential form. The pathogenesis of the secondary forms will depend on the underlying disease.

In cutis verticis gyrata the skin gradually thickens, forming a variable number of folds and furrows that resemble the cerebral cortex. In most cases, the folds are parallel and run in an anteroposterior direction, though they may also be transverse in the occipital region.\(^2-5\) A distinctive feature of this condition is the impossibility of leveling the folds through traction or pressure. The areas affected may differ between individuals; lesions usually develop on the vertex of the scalp, though the temporal, frontal, and occipital regions can occasionally be involved. Accompanying symptoms will depend on the underlying cause of the cutis verticis gyrata, which is a benign disorder that usually requires no treatment. Surgery may be considered if the patient requests treatment for cosmetic reasons. The type of surgery will depend on severity, site of the lesion, and the underlying disease and can include simple excision, the placement of skin expanders, or skin grafts.\(^6\)

References


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